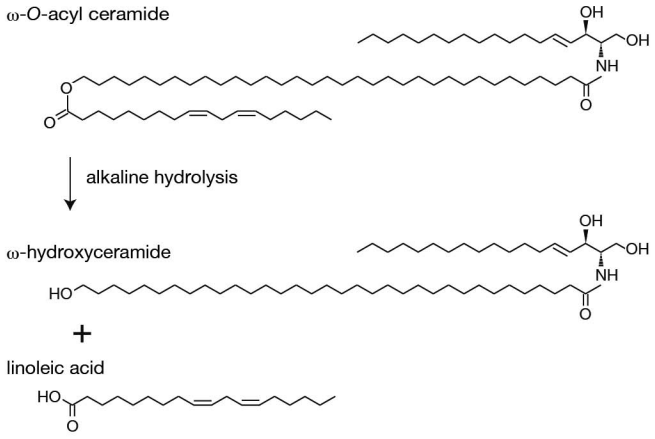


**FIG S1** Structures of ω-*O*-acyl Cer and its breakdown products by alkaline hydrolysis. Schematic representation of the epidermis-specific Cer, ω-*O*-acyl Cer. ω-*O*-Acyl Cer is hydrolyzed to ω-hydroxyCer and linoleic acid by alkaline treatment.

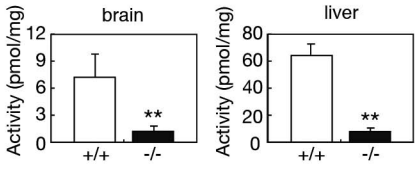


**FIG S2** Decreased FA elongase activity toward C22:0-CoA in brain and liver of *Elovl1* knockout mice. Total membrane proteins prepared from brain (75 μg) and liver (40 μg) of *Elovl1*$^{+/+}$ and *Elovl1*$^{-/-}$ mice were subjected to an *in vitro* FA elongase assay using 50 μM C22:0-CoA and 0.075 μCi [$^{14}$C]malonyl-CoA as substrates. After a 30 min incubation at 37 ˚C, lipids were subjected to methanolysis, extraction, separation by reverse phase TLC, and detection by a BAS-2500 bioimaging analyzer. Values indicate the amounts of FA methyl ester products and represent the means ± S.D. of four independent experiments. Statistically significant differences compared to results for the *Elovl1*$^{+/+}$ mice are indicated (**$p < 0.01$; Student's t-test).

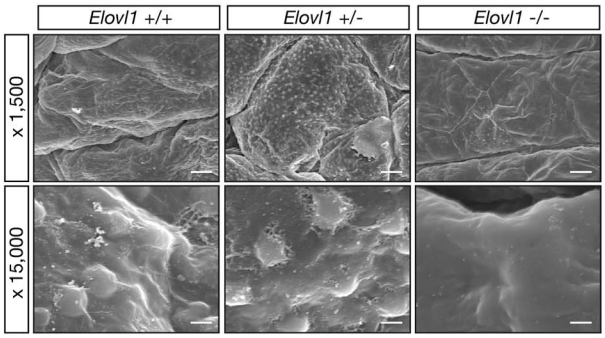


**FIG S3** Abnormally smooth morphology of skin in *Elovl1* knockout mice. Anterior limbs near the shoulder of *Elovl1*$^{+/+}$, *Elovl1*$^{+/-}$, and *Elovl1*$^{-/-}$ mice at E18.5 were subjected to electron microscopy. Magnification factors are indicated at left. Bar, 8 μm (upper panels) and 0.8 μm (lower panels).

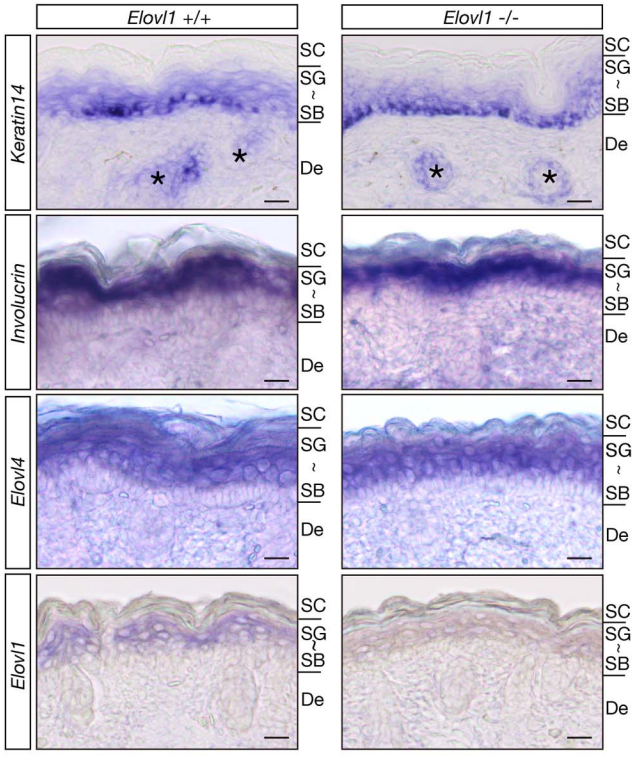


**FIG S4** Normal expression of *Keratin 14, Involucrin,* and *Elovl4* mRNA in skin from *Elovl1* knockout mice. Skin isolated from *Elovl1*$^{+/+}$ and *Elovl1*$^{-/-}$ mice at E18.5 were fixed with 4% paraformaldehyde, hybridized with a digoxygenin-labeled *Keratin14*, *Involucrin*, *Elovl4*, or *Elovl1* RNA probe, and stained with alkaline phosphatase-conjugated anti-digoxygenin antibody ($F_{ab}$ fragment) and nitroblue tetrazolium/5-bromo-4-chloro-3-indolyl phosphate solution. Frozen sections (20 or 25 μm) were subjected to microscopic observation under a DM5000B light microscope and photographed. The signals marked by asterisks indicate the expression of the *Keratin 14* mRNA in the hair follicles. Bar, 50 μm. De, dermis.

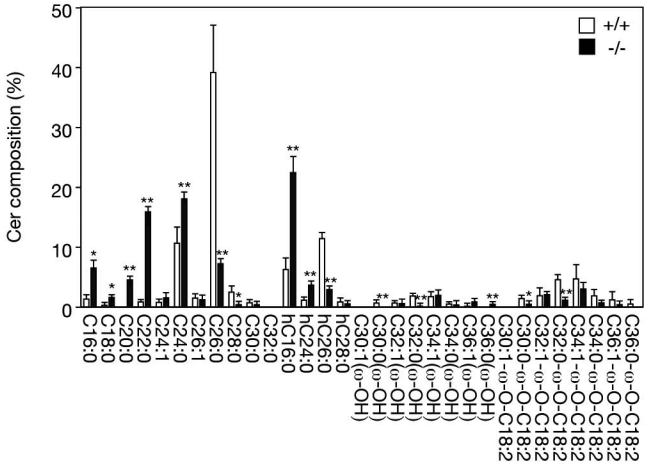


**FIG S5** Decreased ULCFA-containing Cers in epidermis of *Elovl1* knockout mice. Lipids were extracted from the epidermis of *Elovl1*$^{+/+}$ and *Elovl1*$^{-/-}$ mice at E18.5, and subjected to electrospray ionization MS using 4000 QTRAP MS/MS system equipped with the nanoflow ion source TriVersa NanoMate. Cers were identified by precursor ion scan with $m/z$ = 264.4, and analyzed by Analyst software. Values presented are the amount of each Cer species relative to that of total Cers and represent the means ± S.D. from three independent reactions. Statistically significant differences compared to results for the *Elovl1*$^{+/+}$ mice are indicated (*$p < 0.05$, **$p < 0.01$; Student's t-test).

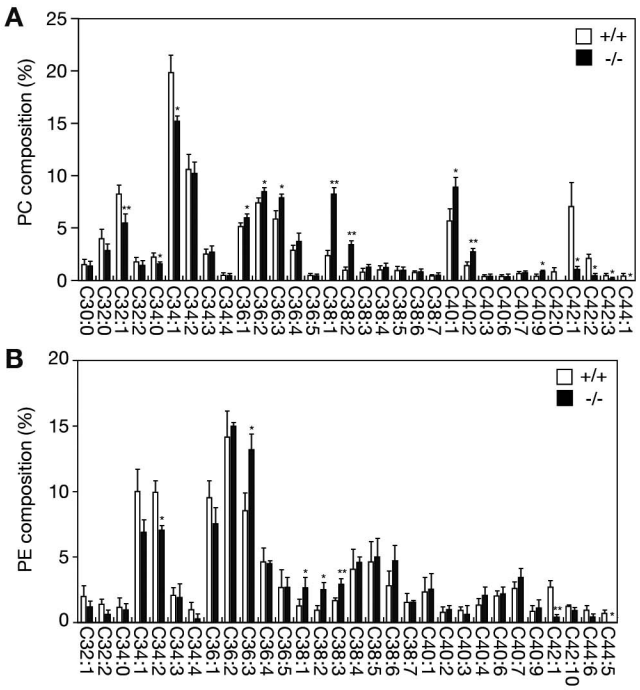


**FIG S6** Changes in PC and PE species in the epidermis of *Elovl1* knockout mice. (A and B) Lipids were extracted from the epidermis of *Elovl1*$^{+/+}$ and *Elovl1*$^{-/-}$ mice at E18.5, and subjected to electrospray ionization MS using 4000 QTRAP MS/MS. PCs (A) and PEs (B) were identified by precursor ion scan ($m/z$ = 184.1) and neutral loss scan ($m/z$ = 141.0), respectively, and analyzed by the automated search engine Lipid Search. Values presented are the amount of each PC or PE species relative to that of total PE or PC, respectively, and represent the means ± S.D. from three independent reactions. Statistically significant differences compared to results for the *Elovl1*$^{+/+}$ mice are indicated (*$p < 0.05$, **$p < 0.01$; Student's t-test).

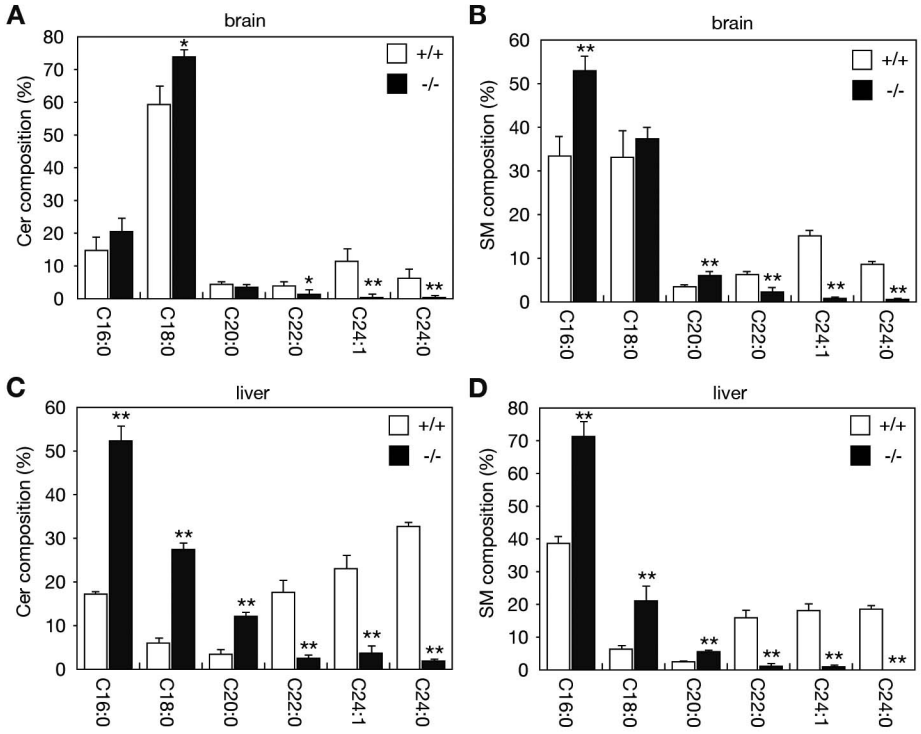


**FIG S7** Reduced levels of C24 Cers and SMs in the brain and liver of *Elovl1* knockout mice. (A-D) Lipids were extracted from brain (A and B) and liver (C and D) of *Elovl1*$^{+/+}$ and *Elovl1*$^{-/-}$ mice at E18.5, treated with alkaline, and subjected to electrospray ionization MS using 4000 QTRAP MS/MS. Cers (A and C) and SMs (B and D) were identified by precursor ion scans (Cer, $m/z$ = 264.4; and SM, $m/z$ = 184.1) and analyzed by Analyst software. Values presented are the amount of each Cer or SM species relative to that of total Cer or SM, respectively, and represent the means ± S.D. from three independent reactions. Statistically significant differences compared to results for the *Elovl1*$^{+/+}$ mice are indicated (*$p < 0.05$, **$p < 0.01$; Student's t-test).